

# Implications of Deafblindness on Visual Assessment Procedures: Considerations for Audiologists, Ophthalmologists, and Interpreters

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Visual assessment of deaf persons presents a challenge to the ophthalmologist and the optometrist. At the time they want to measure visual function, the interpreter uses visual communication, competing for the patient's attention. Important rules of good assessment of visual functioning include taking turns and giving the interpreter sufficient time to convey the questions related to each test. This has implications for audiologists working with this population as well. Communication should be tailored to the varying needs of people who may have long-standing deafness and good sign language, may have lost their hearing at an advanced age and have limited or no sign language, or may be young persons or children who have congenital or progressive loss of hearing combined with congenital or progressive vision loss. Individuals with sudden hearing and vision loss, as well as those with intellectual disabilities and limited language, are particularly challenging groups for the assessment of

vision and require special communication skills for ophthalmologists and audiologists. The ophthalmologist usually knows the nature of vision changes typical of disorders that the patient has. For the clinical follow-up of a condition, a few measurements are usually taken. For a comprehensive assessment of visual functioning for rehabilitation and education, many more measurements, observations, and questions are needed so that vision for communication, orientation in the environment, and performance of near-vision tasks can be reported and the effects of the environment can be evaluated. Testing should cover ocular disorders and vision loss caused by brain damage.

**Keywords:** visual impairment; deafblindness; retinitis pigmentosa; coloboma; night blindness; visual field; contrast sensitivity; visual acuity; ventral stream; dorsal stream

For a comprehensive assessment of visual functioning of deafblind persons for rehabilitation and education, many measurements, observations, and questions are needed (Table 1) so that vision for communication, orientation in the environment, and performance of near-vision tasks can be

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reported and the effects of the environment can be evaluated.<sup>1</sup> Communication during and after an assessment of vision of deaf patients was considered to be an important area to investigate when the Nordic Staff Training Course for Deafblind Services was conceived in 1981. The study was started in 1987 when funding was received from the National Science Foundation for my sabbatical year at the Smith-Kettlewell Eye Research Institute. Subsequently during the winter of 1997-1998, I had the privilege to examine deaf patients with Lindsay Gimble, an unusually skilled interpreter who quickly learned to cope with different kinds of vision problems and varying types of communication. Examinations were recorded on video and were later assessed sentence by sentence. The resulting book, *Assessment of Vision and Hearing of Deaf-Blind Persons*,<sup>2</sup> was published by the Royal

Table 1.

**Assessment of Visual Functioning in Children\***Lea Hyvärinen, MD, [www.lea-test.fi](http://www.lea-test.fi)**OCULOMOTOR FUNCTIONS and CLINICAL MEASUREMENTS**  
Fixation; Following movements; Saccades; Nystagmus

## Strabismus:

## Head position:

## Refraction; Accommodation:

## Grating acuity:

## Visual acuity at near:

Single symbols: binok \_\_\_\_ m/\_\_\_\_ M = \_\_\_\_ RE \_\_\_\_\_ LE \_\_\_\_\_  
 standard near test binok \_\_\_\_\_ RE \_\_\_\_\_ LE \_\_\_\_\_  
 50% crowding test binok \_\_\_\_\_ RE \_\_\_\_\_ LE \_\_\_\_\_  
 25% crowding test binok \_\_\_\_\_ RE \_\_\_\_\_ LE \_\_\_\_\_

## Visual acuity at distance:

single symbols at \_\_\_\_ meter binok \_\_\_\_\_ RE \_\_\_\_\_ LE \_\_\_\_\_  
 line test at \_\_\_\_ meter binok \_\_\_\_\_ RE \_\_\_\_\_ LE \_\_\_\_\_

## Contrast sensitivity:

## Colour vision:

## Motion perception:

## Visual field:

## Visual adaptation to changes in luminance level:

**VISUAL PROCESSING FUNCTIONS:**

## RECOGNITION and READING

## Faces:

## Facial expressions, Body language:

## Landmarks:

## Concrete objects:

## Pictures of concrete objects:

## Geometric forms:

## Letters:

## Numbers:

## Words:

## Crowding effect:

## Reading speed:

## Scanning lines of text:

## Efficiency of reading:

**PERCEPTION OF PICTURES**

## Length of lines:

## Orientation of lines:

## Details of pictures:

## Figure-ground:

## Visual closure:

## Noticing errors:

## Noticing missing details:

## Comparison with pictures in memory:

## 'Reading' series of pictures:

## Visual problems in copying pictures:

**PERCEPTION OF SPACE**

## Depth perception:

## Perception of near space:

## Perception of far space:

## Simultanagnosia:

## Perception of textures and surface qualities:

## Orientation in space:

## Memorizing routes:

## Vision in traffic situations and in playgrounds:

**EYE-HAND COORDINATION**

## Grasping and throwing objects:

## Drawing, free hand:

## Copying, from near/ from blackboard:

## Copying, motor planning and execution:

**INTEGRATION PROBLEMS**

## Vision not used when listening or exploring:

## Vision not used when moving:

## Balance:

**COMPENSATORY STRATEGIES**

## Auditory information:

## Tactile, kinesthetic and haptic information:

## Memory:

**SENSITIVITY TO OVERLOAD****OTHER: EFFECT ON PARTICIPATION, EFFECT OF ENVIRONMENT**

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This list of the most important visual functions to be considered in a comprehensive assessment of visual functioning is used in many rehabilitation services and schools to collect enough information for the planning of services.

Victorian Institute for the Blind in Melbourne, Australia. It sold out several years ago but is now found on my home page (<http://www.lea-test.fi>), which mainly contains information on pediatric vision problems. However, the pages on deafblindness are the pages most frequently visited, representing about 15% to 20% of the half million hits each month. This indicates that there is still a strong demand for information on assessment of the vision of deafblind persons.

Questions related to the assessment of vision can be divided into the following 3 main categories: (1) communication during the assessment, (2) testing visual functions and interpreting test results, and (3) how audiologists should pay attention to vision loss.

**Communication During the Assessment**

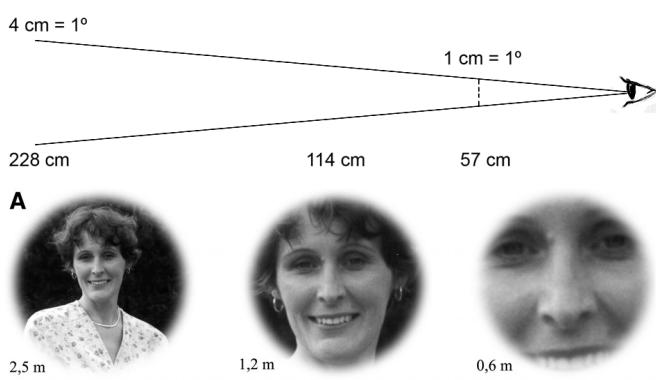
Before the examination of visually impaired, deaf, or hard-of-hearing persons can be executed properly, the ophthalmologist and the interpreter have to learn how to communicate in this rare situation in which vision is being used for communication at the same time as it is being assessed. The patient needs additional information as well. Texts such as "Eyes and Vision" on my home page can be used before the assessment to make it easier for the patient to understand the physician. These easy-to-read texts describe the anatomy and function of eyes and visual pathways, common ocular conditions, and visual functions written at a level for patients to understand.

It is not necessary for the physician to be able to use sign language other than to sign "look here," "look there," "keep looking at this picture," "place your chin here, the forehead against the bar," and a few other expressions often used. However, it is necessary to know enough about the structure of sign language and the deaf culture to be comfortable in this cross-cultural situation. Patients, especially children, become relaxed when they realize that the ophthalmologist knows a few signs and is willing to use them.

The interpreter needs to be familiar with the medical vocabulary. When there is a new term, it is not sufficient to just finger spell it. It needs to be explained by the physician so that it is understandable to the interpreter and to the patient. For example, we measure ocular pressure using applanation tonometry barely touching the cornea, and the sign is usually a fist against the palm of the other hand, which does not describe the gentle way of measuring. Similar details of interpretation may exist in audiology. It would be good to arrange a workshop for studying the signs used in ophthalmology and audiology. One should also study the visibility and readability of speech reading, manual alphabets,<sup>3</sup> and signs used during clinical examinations.

As an example, we will consider retinitis pigmentosa and communication during visual assessment of individuals with this disorder. Retinitis pigmentosa is a common cause of vision loss among the deaf population. It begins to disturb communication when the diameter of the visual field is a few degrees. This is often depicted as "tunnel vision." However, the visual field is not like a tunnel but rather like a cone opening toward the distance (Figure 1).

At 57 cm (approximately 2 feet), 1 cm on a surface is equal to 1° of visual angle. Therefore, if a person's



**Figure 1.** The small central visual field is not a tunnel but a cone that opens toward the distance. This affects communication situations. The closer you go toward a person with a small visual field, in this case approximately  $10^\circ$ , the less you are visible. The small central visual field is seldom as regular and round as in this picture but is often slightly oval horizontally.

visual field is  $10^\circ$  in diameter (a usual size among adult persons with retinitis pigmentosa), the visual field covers a space 10 cm in diameter at 57 cm, 20 cm at 114 cm, and 40 cm at 228 cm (approximately 8 feet). Thus, a distance of 6 to 8 feet would be a good distance for signing.

At the beginning of a communication situation, the interpreter should test the size of the communication field by asking the patient or client to indicate at which point the hands of the interpreter become visible when brought from the side toward the body of the interpreter. When the approaching hand becomes visible, the interpreter touches his or her body at that point to provide a reference of the edge of the field for signing.

Another challenging group is deaf persons developing macular degeneration. When central vision is affected, lip-reading and finger spelling are not perceived, and signs have to be slower than usual and often repeated. If vision loss is severe, an elderly patient may need to have a family member or a friend to function as a relay interpreter (Figure 2).

Offices of ophthalmologists and optometrists are often dark, with focal illumination on the test charts. Such illumination is not acceptable when testing visual functioning of a deaf person. This will also affect communication with an audiologist and an interpreter when there is poor illumination in a test booth. The interpreter needs to be well seen by the patient and an optimal distance chosen for the use of sign language. Therefore, before the assessment starts, the size of the communication field and

the visibility of finger spelling and lip movements (if speech reading is used) need to be tested. Certain test situations such as Goldmann perimetry to measure visual field require the use of tactile communication, which needs to be explained to the patient before each test.

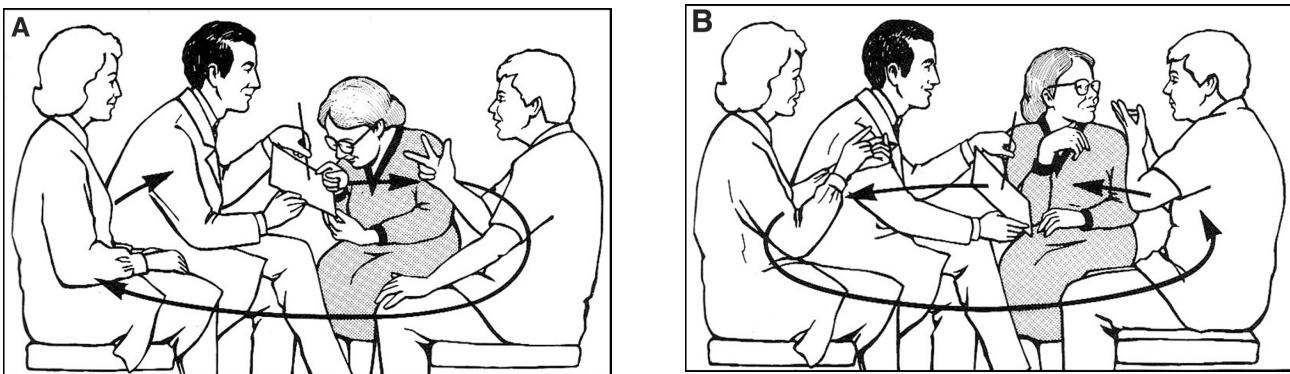
## Testing Visual Functions and Interpreting Test Results

The most common examination measurement, visual acuity, is easiest to measure using a standard number chart on a light box. Numbers are easier to read than letters in finger spelling; even an ophthalmologist can recognize them. Young children prefer the Lea Symbols, pictures of concrete objects, to the much more abstract letters. Contrast sensitivity measurements take almost no time when they are measured using visual acuity charts at low contrast levels.

For color vision, the Waggoner test (similar to the Ishihara test) works well for children and with adults in screening for inherited regular color deficiencies. The Lea Panel 16 can be used for quantitative measurements. It is a modification of the Farnsworth Panel D-15 with large test stimuli, which can be reduced to the standard size by placing gray rings on the color surface.

Visual adaptation to lower luminance levels can be assessed as the cone adaptation time using the Cone Adaptation Test (Good-Lite). It was designed for early screening of retinitis pigmentosa among deaf children and functions well in the screening of children from the age of 3 years. It depicts functioning better than the expensive electroretinography test because the parents can watch their child's performance at different luminance levels and understand the need for a flashlight if the child cannot see the test objects at low luminance levels.

In all the tests discussed so far, testing and interpretation of the test results are easy. The clinical gold standard, measurement of visual field using the Goldmann perimeter, causes many problems. Visual communication during the test is impossible because the patient is sitting facing the cupola of the perimeter and sees only the white test surface in front of him or her. Therefore, the test situation needs to be explained to the patient in detail before the test starts. Children understand the test situation best if they can watch an older child being tested. Older children seem to be able to describe



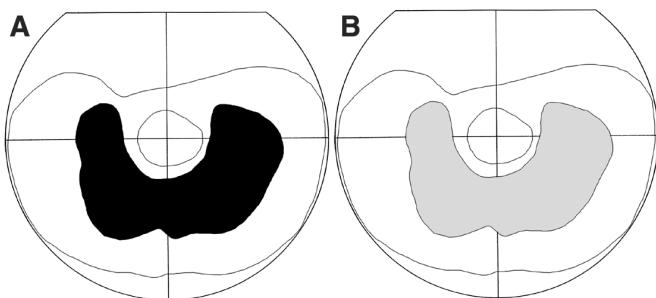
**Figure 2.** Relay interpreting. If a person cannot read the signs of the interpreter, a family member or a friend can function as a relay interpreter. The information then goes through 2 interpreters, which takes time. If the relay interpreter is a hearing person, the interpreter is still needed to check that the information transfer is correct.

the task to younger ones so well that it would be worthwhile to video record these explanations for training of interpreters. To keep the communication going during the test, it is advisable to agree with the patient that tactile information will be used, for example a tap on the knee, if fixation starts to wander off from the central fixation mark.

In a comprehensive assessment for rehabilitation and education, many more tests are needed than can be covered in this short review (Table 1). Many observations require participation of all members of the rehabilitation team, family members, therapists, and teachers.

Interpretation of the test results requires experience, and interpretation of Goldmann perimetry results is especially difficult. The measurement is performed at a low photopic level, 10 to 15 candela/m<sup>2</sup>, which is at the border between photopic (daylight) and mesopic (twilight) vision. Scotomas, which are areas of loss or impairment of sensitivity surrounded by a field of normal vision, measured at that low luminance level are larger than they are in room light and are much larger than they are in daylight.

The implications of measurement at low luminance levels can be demonstrated with the following case. More than 10 years ago, a young man with Usher syndrome was found to have large absolute scotomas in the lower part of both visual fields (Figure 3A). It was time for career planning, so I was asked for a second opinion on the finding. I had assessed the visual fields of this young man ever since he was 4 years old and knew that he was the



**Figure 3.** A, Absolute scotoma, an area of loss of sensitivity, in the lower part of the visual field of a young ice hockey goalkeeper. B, With flicker sensitivity, there was measurable function in the area of the absolute scotoma. Therefore, the scotoma area has been painted gray, depicting the relative scotoma.

best goalkeeper in the local youth ice hockey league. I thought that there had been some problems in communication during the measurement and suggested that I measure the Goldmann fields once more. Figure 3A. is drawn based on my measurement placing the visual fields of both eyes in one picture. There was a large absolute scotoma in the lower visual fields of both eyes. Because the young man was so exceptionally good at ice hockey, I arranged for the measurement of flicker sensitivity (sensitivity to flickering stimuli instead of the nonflickering stimuli in Goldmann perimetry) in the area of the scotoma. At each point, there was measurable flicker sensitivity. The scotoma was blind to the slowly moving dot in the Goldmann perimetry, but there was still motion perception in that scotomatous area. The magnocellular

pathway that carries movement-related information collects information from large areas in the retina and can transfer enough information for motion perception when small forms cannot be perceived.

Since then, absolute scotomas on Goldmann perimetry have been shown to contain useful vision at higher luminance levels and for motion perception in several young patients with retinitis pigmentosa. Similarly, in hemianopia that is caused by a lesion in the posterior part of the optic radiation, there may be an absolute scotoma with or without macular sparing on Goldmann perimetry but measurable activity on flicker perimetry. With training, flicker sensitivity may improve to the same level as in the normal half field.<sup>4-9</sup> This represents a new challenge in vision rehabilitation.

Motion perception is not examined during routine clinical assessments. It may decrease in diffuse retinal degenerations such as retinitis pigmentosa and in local lesions of brain cortex. This has significant implications for audiologists who consider the communication function of their clients. If motion perception is poor, lip movements and finger spelling become blurred and may disappear. Most often, there is loss of rapid movements, but slow movements may still be perceived (measurable using the Pepi-test, available for copying at <http://www.lea-test.fi>). In these cases, finger spelling can be read if the signer makes a short stop between each letter. Lip movements are more difficult to modify. However, it is advisable to test how slow the lip movements should be so that they do not become blurred. This can be done by using video films at varying speeds. Some children with impaired perception of high-speed movements experience the blurring or disappearance of lips when a person talks to be so unpleasant that they turn their heads away. This has sometimes been a problem in speech therapy if the therapist has not known why the child refuses to look at the therapist's mouth for correct form and placement of the tongue and lips. Head turning has also been interpreted as an autistic behavior.

Loss of motion perception also affects moving. When we move, objects in our surrounding are seen moving in the opposite direction with varying angular speed depending on the distance from us. This gives us important information about the structure of the environment. If motion perception is defective, objects in the side view may be perceived as blurred, which results in the person walking fast. Then the blur disappears because the objects disappear from the visual field of the person. Therefore, the fast-walking person may bump into large objects, despite

good central vision. This behavior is often not understood by the person or by people around him or her. The phenomenon is also difficult to explain to the person. A short lecture on visual pathways and brain cortex is necessary for creation of a vocabulary that is meaningful to the interpreter and the patient.

## **How Audiologists Should Pay Attention to Vision Loss**

The number of patients with brain damage that affects processing of auditory and visual information is growing in all Western countries and in large cities of developing countries. This is because of improved prenatal and postnatal care of small prematurely born infants, infants with malformations, and children and adults who have had circulatory failure, radiation damage, or accidents affecting brain function. These patients have varying clinical findings that are often difficult to interpret. Adults who have seen and heard normally can often describe what is missing or distorted in their hearing and vision. Young children and school-age children are a challenge in communication because to them their way of hearing and seeing is the only way they know. Clinical neuropsychological tests cover only a part of more than 30 visual functions that need to be assessed by the rehabilitation team together with the child's therapist, teachers, and parents. It takes all the preschool years and several school-age years before the functional situation is understood and rehabilitation and special education programs fit the needs of each child.

When a severely visually impaired person (who is accustomed to depend on hearing for orientation in space) loses hearing in one ear, he or she becomes functionally deafblind in spatial orientation. This sometimes is not appreciated in audiology departments if the personnel is unfamiliar with problems related to vision loss. Patients have sometimes been sent back with a report that their hearing loss does not affect communication, although the referral was for auditory orientation in the environment. Other audiology departments are helpful in rehabilitation, fitting hearing aids and providing listening training. Some hospitals do not accept loss of hearing in one ear as a cause of dual sensory disability if hearing in the other ear is normal or close to normal even if the person is severely visually impaired. This is an area in rehabilitation in which cooperation between ophthalmology and audiology should be improved.

If a visually impaired child has hearing in only one ear, this should be considered in his or her

placement in the classroom, with the hearing ear toward the group of children and placement as close to the teacher as necessary because of the impaired vision. The same rule should be followed during meetings as well.

When a hearing-impaired person has visual impairment, it is important to consider whether the 2 impairments together cause disability in any functional areas defined by the *International Classification of Functioning, Disabilities and Health*<sup>1</sup> for adults and by the *Management of Low Vision in Children*<sup>10</sup> for children. In some eye hospitals, the *International Classification of Functioning, Disabilities and Health* is not yet followed in assessments for rehabilitation and special education. A hearing-impaired person's communication may be affected by changes in motion perception, contrast sensitivity, or visual field even at a time when visual acuity is still normal or near normal. Similarly, many other functional areas may be affected if visual acuity and the size of the visual field are normal, for example moving in twilight conditions on the way to school or work if adaptation to low luminance levels has been lost.

Communication problems during assessment by audiologists are similar to those in offices of ophthalmologists. The position of the interpreter should be chosen correctly, the luminance level should fit the needs of the patient, and the patient should be asked whether the interpreter has had time to check the communication distance and the size of the communication field at that distance, whether the luminance level is correct, and whether there is anything else that the physician should be aware of such as whether the patient uses speech reading. If so, it should be determined whether he or she could read the lip movements or whether they should be slower.

Working with a good interpreter makes assessment of visual functioning and assessment of hearing possible even in complicated cases and when communication is limited. Training of interpreters

in the management of vision problems is recommended. Interpreting in ophthalmology and audiology settings will be more relaxed and more exact when deaf culture and sign language are familiar to the ophthalmologist, changes in vision affecting communication are known to the audiologist, and there is good interpretation. A follow-up visit of a deafblind patient is often experienced as an enjoyable part of the day and a pleasant challenge.

## References

1. World Health Organization. *International Classification of Functioning, Disabilities and Health*. Geneva, Switzerland: World Health Organization; 2001.
2. Hyvärinen L, Gimble L, Sorri M. *Assessment of Vision and Hearing of Deaf-Blind Persons*. Melbourne, Australia: Royal Victorian Institute for the Blind; 1988.
3. Hyvärinen L, Gimble L, Sorri M. Tests, assessment, education. <http://www.lea-test.fi>. Accessed March 24, 2007.
4. Raninen AN, Näsänen RE, Hyvärinen L. Detecting flicker and discriminating letters in the blind hemifield [abstract]. *Invest Ophthalmol Vis Sci*. 1998;39:399.
5. Raninen AN, Näsänen RE, Hyvärinen L. Rehabilitation of vision in the hemianopic field [abstract]. *Invest Ophthalmol Vis Sci*. 1999;40:737.
6. Vanni S, Raninen A, Näsänen R, Tanskanen T, Hyvärinen L. Dynamics of cortical activation in a hemianopic patient. *Neuroreport*. 2001;26:12(4): 861-865.
7. Hyvärinen L, Raninen AN, Näsänen RE. Vision rehabilitation in homonymous hemianopia. *Neuroophthalmology*. 2002; 27(1-3):97-102.
8. Raninen A, Vanni L, Hyvärinen L, Näsänen R. Temporal sensitivity in a hemianopic visual field can be improved by long-term training using flicker stimulation. *J Neurol Neurosurg Psychiatry*. 2007;78(1):66-73.
9. Henriksson L, Raninen A, Näsänen R, Hyvärinen L, Vanni S. Training-induced cortical representation of a hemianopic hemifield. *J Neurol Neurosurg Psychiatry*. 2007;78(1):74-81.
10. *Management of Low Vision in Children*. Geneva, Switzerland: World Health Organization; 1993.